Original articles

Quality of life in patients with cystic fibrosis and their parents: what is important besides disease severity?

D Staab, K Wenninger, N Gebert, K Rupprath, S Bisson, M Trettin, K D Paul, K M Keller, U Wahn

Abstract

***Background*—Cystic fibrosis is the most common inherited disease with a fatal outcome in industrialised nations. With the improvement in life expectancy, supporting patients and their families in adapting to life with this chronic progressive disease has become increasingly important. The aim of the present study was to investigate the relationship between health related quality of life (HRQOL) in this population, severity of disease, and cognitive/behavioural factors such as subjective health perception and ways of coping.**

***Methods*—A sample of 89 adolescent and adult patients with cystic fibrosis and 125 parents of younger patients with cystic fibrosis completed questionnaires on health related quality of life and on ways of coping with the illness. Parents were asked to fill out the questionnaires regarding their own quality of life and coping. Multiple regression analyses were performed to examine the relationship between different predictor variables and quality of life.**

***Results*—After accounting for the impact of disease severity and hours of treatment per day, the subjective health perception of patients significantly explained variance in their quality of life. Ways of coping were also significantly correlated with HRQOL. In parents the most important factor in explaining variance of HRQOL seems to be the coping style, whereas disease severity of the child and subjective health perception did not show any influence.**

***Conclusions*—The findings support the important role of cognitive and behavioural factors in specific subjective health perception and ways of coping in the adaptation to this severe chronic disease, both in patients themselves and in parents. The results call for a careful assessment of issues of coping and professional support for families of patients with cystic fibrosis in the early course of disease.**

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Cystic fibrosis is the most common inherited disease with a fatal outcome in industrialised nations, and is becoming increasingly prevalent as survival is prolonged. Thirty years ago most of these patients died in infancy but today about 36% reach adulthood.[1] On the basis of epidemiological analysis we can expect children born with cystic fibrosis in 1990 to have a life expectancy of 40 years—in other words, more than 90% will reach adulthood.[2] This will entail costly and time consuming therapy. As in other chronic pulmonary diseases, lung function parameters such as forced expiratory volume in one second (FEV_1) are poor predictors of the degree of disability.[3 4] Thus, apart from purely somatic factors such as lung function, weight for height, or clinical scores like the Shwachman score, psychosocial aspects of living with this chronic fatal disease may be important additional outcome variables in clinical studies. They have increasingly become a focus of attention in efforts to achieve the best possible outcome for the individual patient.

The goal of the present study was to investigate the relationship between health related quality of life (HRQOL) and a number of medical and psychosocial variables in a population of adolescent and adult patients with cystic fibrosis and parents of younger patients. Studies on HRQOL in patients with cystic fibrosis have usually focused on its relationship with clinical status alone.[5–10] Most of these studies found only moderate correlations with somatic parameters. The aim of the present study was to extend this research by investigating whether other factors such as subjective perception of health and ways of coping with disease are related to HRQOL in cystic fibrosis. To our knowledge associations between HRQOL and these psychosocial factors have not yet been investigated.

HRQOL, a construct to describe the state of well being, consists of two components: (1) the ability to perform everyday activities which reflect physical, psychological, and social well being, and (2) satisfaction with levels of

functioning and the control of disease and/or treatment related symptoms.[11]

There is no clear agreement on whether to use generic or disease specific instruments in the assessment of HRQOL. While specific instruments focus on problems associated with single disease states, generic measures apply to a variety of health states and diseases and allow for broad comparisons. Since we were interested in the HRQOL of parents as well as of patients, a generic instrument, the German questionnaire "*Alltagsleben*" ("*Every Day Life*")[12] which also applies to the general population, was chosen. Its reliability and validity has been confirmed in studies with different disease groups and healthy volunteers. A disease specific instrument was not available at the time of the present study. Recently, a specific measure has been developed and is currently under evaluation.

The results presented in this paper are part of the baseline study in the evaluation of an educational programme for patients with cystic fibrosis and their families. Our main interest was to evaluate which factors besides disease severity are responsible for the variance in HRQOL.

Methods

SUBJECTS

Two samples of 89 adolescent and adult patients with cystic fibrosis and 125 parents of children with cystic fibrosis were recruited from four German Cystic Fibrosis outpatient clinics in Bonn, Dresden, Berlin-Buch and Berlin-Heckeshorn, about 80% of all patients with cystic fibrosis seen at these outpatient clinics at that time. (There was a small cross-over of 16 patients from the patient sample whose parents participated in the parent sample. Data of both samples were analysed separately). A problem in understanding the German language was the only exclusion criterion.

ASSESSMENT INSTRUMENTS

Subjects completed questionnaires on socio-demographic variables, quality of life, and ways of coping with chronic illness. In addition, the subjects' subjective perception of their own or their child's health was assessed on a five point rating scale covering health states from "little impaired" to "very much impaired". Medical information was taken from the medical chart.

The total score of the questionnaire "*Alltagsleben*" ("*Every Day Life*")[12] which was developed for German speaking populations was used to assess HRQOL. The "*Alltagsleben*" is a 42 item generic instrument with six subscales assessing physical, emotional, social and functional components of quality of life, as well as joy of life and satisfaction with medical care. The total score is composed of all 42 items. Since all six subscales were strongly correlated with the total scale (patients: $r = 0.58–0.92$, $p<0.001$; parents: $r = 0.60–0.93$, $p<0.001$), the total score was used as an outcome measure providing a summary of overall HRQOL. The reliability and validity of the questionnaire were supported in a number of studies including healthy subjects as well as patients with various chronic diseases.[12]

The German 102 item questionnaire "*Freiburger Fragebogen zur Krankheitsverarbeitung*" (FKV) ("*Freiburg Questionnaire of Coping with Disease*")[13] was used to assess different ways of coping with illness. The concept that is measured by this questionnaire can be defined as the emotional, cognitive, and behavioural efforts of an individual to master or manage the existing or expected demands of a disease.[13] The questionnaire is composed of the following 12 subscales: problem solving (item example: "I came up with a plan for what I have to do and acted accordingly"), depressive coping (for example, "I asked myself again and again why me"/"I resigned"), hedonism (for example, "I decided to try to enjoy life as much as possible"), religiousness/search for meaning (for example, "I praised"/"I tried to see the disease as a chance"), distrust/pessimism (for example, "I was distrustful whether anything would be missed or overlooked"/"I was prepared for the worst"), cognitive avoidance (for example, "I refused to take this seriously"), distraction/self-confidence (e.g. "I told myself that there were worse things that I had coped with before"), control of emotions/retreat from social relations (for example, "I tried to keep my feelings to myself"), regressive tendency (for example, "I wished I was allowed to be weak"), social comparison (looking at your own situation in relative terms, for example, "I told myself that others were worse off"), compliance/trust in treatment (for example, "I did exactly what my doctor advised me to do"), and self-encouragement (for example, "I was convinced that the treatment would be successful"). The reliability and validity of the FKV have been supported in studies involving patients with various chronic diseases. Parents were instructed to answer the questionnaire with regard to how they were coping with the disease of their child.

PROCEDURE

The study was conducted between June 1993 and August 1994. Questionnaires were filled out at a clinic visit and took about 30 minutes to complete. All questionnaires were self administered; both patients and parents filled out the same questionnaires. Patients in the Berlin centres were offered an opportunity to participate in an educational programme for families of cystic fibrosis sufferers. Support services available to all subjects included individual counselling by a psychologist.

STATISTICAL ANALYSES

The Statistical Package for the Social Sciences (SPSS for Windows) was used for data management and analyses.

Multiple regression analyses were performed to examine the relationship between various predictor variables and quality of life. A hierarchical procedure was used entering medical variables into the equations in a first block. This procedure was chosen to identify the additional variance explained by subsequently

Table 1 Clinical data of patients with cystic fibrosis (CF)

	CF patients (n = 89)			*CF children of parents questioned (n = 125)*		
	Mean	*Range*	*SD*	*Mean*	*Range*	*SD*
Shwachman score	57.2	15–75	12.7	61.8	20–75	12.4
% of ideal body weight	95.5	60–166	14.8	95.6	72–126	9.8
FEV_1*	55.6	16–118	24.7	74.1	28–118	22.5
	Percentage of sample			Percentage of sample		
Pseudomonas colonisation	66.3			44.0		
Cirrhosis of the liver	11.2			5.6		
Insulin dependent diabetes	14.6			0.8		

*FEV_1 was only obtained from children over the age of five which reduces the number of children in the parent sample from 125 to 95 for this variable.

entered variables beyond what is already explained by medical scores.

The following variables were used as predictors in the regression models: Shwachman score, percentage of ideal body weight, FEV_1, hours spent on treatment per day, subjective health perception rating, and a number of subscales of the ways of coping questionnaire. Of the latter, only scales that were related to quality of life (r with $p<0.10$) but were not highly intercorrelated ($r<0.45$) were used in order to reduce the overall number of predictors entered into the equation. A listwise deletion of missing data led to slightly different numbers of subjects being included in different analyses.

Results

DESCRIPTION OF THE SAMPLES

The samples recruited in the four different outpatient clinics did not differ in socioeconomic status nor in the dependent variable HRQOL. They were therefore regarded as one group for statistical analyses.

Forty male and 49 female patients with cystic fibrosis of mean (SD) age 24.2 (6.9) years (range 12–49) participated in the study. Of these, 32% were at school or in vocational training, 38% were working, 9% were unemployed, and 15% were receiving a disability pension. About half of the sample (49%) were single and living with their parents, 16% were living alone, and 30% were living with a partner or spouse. As shown in table 1, the patients' clinical scores indicated a wide range of disease severity.

As a second sample, 125 parents of patients with cystic fibrosis participated in the study. One parent per patient was asked to fill out the questionnaires—namely, the primary care provider to the child with cystic fibrosis—resulting in a sample of 111 mothers and 14 fathers. HRQOL scores of fathers did not differ significantly from those of the mothers so that they were regarded as one group for statistical analyses. Being a single parent or having more than one child with cystic fibrosis did not impact on parents' HRQOL scores. The mean (SD) age of the patients was 9.6 (4.9) years (range 1–22). At the time of the study all patients in this group were living with their parents. Their clinical scores are shown in table 1.

In both samples, the total quality of life score was not significantly correlated with socioeconomic status as reflected by an index composed of income and educational status.

RELATION BETWEEN DISEASE SEVERITY AND SUBJECTIVE HEALTH PERCEPTION

In both samples the subjective health perception rating and disease severity, as indicated by the Shwachman score,[14] were moderately correlated. For patients the Pearson correlation coefficient was –0.45 ($p<0.001$). Lower scores on the subjective rating scale represented better health whereas lower Shwachman scores indicated greater disease severity thus resulting in a negative correlation. Dividing both variables into three categories of slightly, moderately and severely impaired health, three patients (3.4%) with a low Shwachman score viewed themselves as only slightly impaired while three patients with a high Shwachman score rated their health as severely impaired—that is, 6.8% of the sample exhibited extreme divergence on the two variables. For parents the Pearson correlation coefficient between disease severity and subjective perception of their child's health was –0.43 ($p< 0.001$); 8.2% of the sample showed extreme divergence between subjective health perception and objective health status.

PREDICTING PATIENTS' QUALITY OF LIFE

The total HRQOL scores for patients were normally distributed with a mean (SD) of 4.0 (0.6) on a five point scale, higher scores representing better HRQOL.

A multiple regression analysis was conducted to test whether patients' subjective perception of their health had additional explanatory value for the quality of their life after accounting for the impact of disease severity and hours spent on therapy. The Shwachman score, percentage of ideal body weight, and FEV_1 were entered into the equation as predictors in block 1, number of hours spent on therapy per day was entered in block 2, and the patients' subjective ratings of their health was added in block 3. Overall, 31% of the variance in quality of life was shared by the predictor variables, the medical scores explained 14% of the variance, while hours of therapy added another 11%. After accounting for the impact of disease severity and hours of therapy as an objective measure of therapy constraints, the patients' subjective perception of their health still had additional explanatory value for differences in quality of life (another 6% of explained variance). Pearson correlations indicated that the subjective health perception was the strongest predictor of patients' quality of life. Results are shown in table 2.

In a second multiple regression analysis (table 3) the relation between ways of coping and quality of life was explored. Three scales of the ways of coping questionnaire were chosen as predictors because of their significant correlation with quality of life and their low intercorrelations—depressive coping, cognitive avoidance, and social comparison. After accounting for the impact of disease severity, ways of coping accounted for an additional 12% of the variance in quality of life. Pearson correlations indicated that social comparison was the strongest predictor. Social comparison was related to higher quality of life whereas

depressive coping was negatively related to quality of life.

PREDICTING PARENTS' QUALITY OF LIFE

The total HRQOL scores for parents were normally distributed with a mean (SD) of 3.8 (0.7) on a five point scale.

A multiple regression analysis with Shwachman score and percentage of ideal body weight entered as predictors in the first block revealed that parents' quality of life was unrelated to disease severity. However, hours spent on daily therapy had a significant impact (7% explained variance). The parents' subjective perception of their child's health did not significantly add to the prediction. The results are displayed in table 2.

While the set of predictors from the first multiple regression analysis had only little explanatory value for parents' quality of life, ways of coping with their child's disease explained 26% of the variance in a second multiple regression analysis (table 3). Five scales of the ways of coping questionnaire were entered into the equation as predictors: depressive coping, social comparison, control of emotions/retreat from social relations, compliance/trust in treatment, and self encouragement. Independent of disease severity, depressive coping and control of emotions/retreat from social relations were negatively related to quality of life. These two scales were the most important predictors with the highest Pearson correlations. Higher compliance/trust in treatment and self encouragement were significantly related to better quality of life.

Discussion

In *patients* with cystic fibrosis subjective perception of health and ways of coping explained a significant amount of variance of HRQOL, beyond what was accounted for by disease severity and time spent on therapy. A depressive way of coping was negatively related to HRQOL. Social comparison—that is, looking at one's own situation relative to others—was related to better HRQOL. It is noteworthy that the majority of patients perceived their own health status realistically, as indicated by good convergence of their subjective health rating with the Shwachman score. However, in cases where patients perceived their health as bad even though a high Shwachman score indicated little impairment, patients reported low HRQOL and vice versa. This underlines the importance of taking subjective cognitive factors into account when discussing issues such as health status, prognosis, or motivation

Table 2 Subjective health perception and health related quality of life (HRQOL): hierarchical regression analyses I

	Patients (n = 83)			*Parents (n = 113)*		
Predictors	*r with HRQOL*	*Explained variance*	*β (with all predictors in equation)*	*r with HRQOL*	*Explained variance*	*β (with all predictors in equation)*
Block 1: Medical scores						
Shwachman score	0.40***		0.06	0.08		0.02
Percentage of ideal body weight	0.11		0.05	–0.07		–0.09
Forced expiratory volume (FEV_1)	0.31**		0.12	Not entered		Not entered
		Adj R^2 = 0.14**			Adj R^2 = 0.00	
Block 2:						
Hours spent on therapy per day	–0.45***		–0.29**	–0.30**		–0.26*
		Adj R^2 = 0.25***			Adj R^2 = 0.07*	
		Adj R^2_{change} = 0.11***			Adj R^2_{change} = 0.07**	
Block 3:						
Subjective perception of health	–0.48***		–0.31**	–0.19*		–0.08
		Adj R^2 = 0.31***			Adj R^2 = 0.07*	
		Adj R^2_{change} = 0.06**			Adj R^2_{change} = 0.00	

*p <0.05; **p <0.01; ***p <0.001.
r = Pearson correlation coefficient; Adj R^2 = adjusted squared multiple correlation (amount of explained variance), adjustment is made for sample size; Adj R^2_{change} = increase in adjusted squared multiple correlation; β = standardised regression coefficient (weight by which predictor is multiplied in the equation).

Table 3 Ways of coping and health related quality of life (HRQOL): hierarchical regression analyses II

	Patients (n = 84)			*Parents (n = 112)*		
Predictors	*r with HRQOL*	*Explained variance*	*β (with all predictors in equation)*	*r with HRQOL*	*Explained variance*	*β (with all predictors in equation)*
Block 1: Medical scores						
Shwachman score	0.35**		0.26*	0.12		0.13
Percentage of ideal body weight	0.10		–0.11	–0.06		–0.17
Forced expiratory volume (FEV_1)	0.36***		0.24*	Not entered		Not entered
		Adj R^2 = 0.14**			Adj R^2 = 0.02	
Block 2: Ways of coping						
Depressive coping	–0.27**		–0.23*	–0.27**		–0.31**
Cognitive avoidance	–0.20*		–0.12	Not entered		Not entered
Social comparison	0.19*		0.31**	0.14		0.08
Control of emotions/retreat from social relations	Not entered		Not entered	–0.37***		–0.30***
Compliance/trust in treatment	Not entered		Not entered	0.18*		0.23*
Self encouragement	Not entered		Not entered	0.22*		0.22*
		Adj R^2 = 0.26***			Adj R^2 = 0.27***	
		Adj R^2_{change} = 0.12**			Adj R^2_{change} = 0.26***	

*p <0.05; **p <0.01; ***p <0.001.
r = Pearson correlation coefficient; Adj R^2 = adjusted squared multiple correlation (amount of explained variance), adjustment is made for sample size; Adj R^2_{change} = increase in adjusted squared multiple correlation; β = standardised regression coefficient (weight by which predictor is multiplied in the equation).

for treatment adherence with the individual patient.

For *parents* the situation was somewhat different. In contrast to patients, parents' HRQOL was not affected by the severity of the disease of their child. However, a large proportion of the variance in HRQOL could be explained by ways of coping. One possible explanation for these results is that choosing certain strategies of dealing with the illness enables parents of even a severely ill child to adapt to the situation and maintain their quality of life. While "depressive coping" and "control of emotions/retreat from social relations" were negatively related to HRQOL, "compliance/trust in treatment" and "self encouragement" seemed to be helpful strategies showing a positive association with HRQOL. Thus, it seems especially important for parents to be able to express their emotions and call on social support, in addition to finding medical care that they can trust for their child. The finding that higher trust in treatment and compliance was significantly related to higher HRQOL in parents suggests that a cooperative relationship between medical staff and parents is also an important factor for parents' HRQOL.

Overall, our findings indicate that cognitive and behavioural factors such as health perception and ways of coping play an important part in HRQOL in patients with cystic fibrosis and their families. However, since our results are cross sectional, they do not allow for causal interpretation. There are several possible reasons for the relations found. Firstly, a depressive way of coping may lead to a lower reported quality of life because the perception of the situation by the patients and parents is dominated by feelings of resignation, helplessness, and sadness. On the other hand, a low overall quality of life may lead to depressive symptoms such as resignation. Longitudinal studies are needed to investigate the causal direction of the association between coping and HRQOL.

In the present study the parent sample comprised mostly parents of school aged patients with cystic fibrosis. Thus, the generalisability of the results to parents of older adult patients is limited. This age group has to cope with different issues from younger patients such as sexual identity, transition into adult care, and employment.

Studies on HRQOL in patients with cystic fibrosis have usually focused on its relationship with clinical status alone.[5–10] Most of the studies found only moderate correlations with somatic parameters such as lung function. Thus, it was speculated that other factors must influence HRQOL in cystic fibrosis. To our knowledge, correlations with ways of coping with the disease have not been investigated previously. The findings of the present study give a first indication of the way in which coping is positively related to HRQOL, and thus may be helpful in dealing with the disease and in deciding which coping strategies seem to be maladaptive. Coping style is one possible target variable in psychological interventions for patients and families with cystic fibrosis—for instance, methods of cognitive therapy and patient empowerment may be helpful in preventing an unfavourable depressive coping style.

The results of this study indicate the need for psychological support around issues of adapting to and coping with this severe and chronic disease, in addition to providing medical care.

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